328 MBL serum levels in children with CF-associated liver disease

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Background: Genetic modifiers like mannose binding lectin (MBL2) gene influence occurrence and severity of CF associated liver disease (CFLD). Variants of MBL genotypes encode low serum levels of MBL protein.
Aim study was to assess MBL serum levels in CFLD patients (pts), comparing to CF pts without liver disease.
Methods: Study was performed in Pediatric II Department and National Cystic Fibrosis Centre Romania, during May-September 2007. 35 CF children aged 2–18 years, presented to annual assessment were evaluated. We consider 3 groups of patients, as follows: group 1 (21 CFLD patients), group 2 (11 pts CF no CFLD), group 3 (15 controls). Groups were age matched. Patients associating diseases who influence MBL level were excluded. MBL assay procedure was performed using MBL Oligomer ELISA kit. Data were statistically analyzed with ANOVA.
Results: 46% CFLD pts were MBL deficient. Among CFLD pts (group 1) MBL average was 2061.99 ng/ml, half of control's average ($p = 0.001$). Group 2 had an average lower with 13% comparing to controls.
Conclusions: Lower MBL average in CFLD patients sustains that MBL deficiency is associated factor for CFLD. Increased MBL levels in CFLD patients could be resultant of increase growth hormone levels in childhood or predominance of "wild" MBL2 genotype in our patients. Longitudinal evaluation of MBL levels and MBL2 genotyping would be useful to identify the patients predisposed to develop CFLD.

329 The pancreas is not visible as an entity by magnetic resonance imaging in the majority of cystic fibrosis patients

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Background: Our aim was to study the volume of the pancreas in adult cystic fibrosis (CF) patients with and without CF related diabetes (CFRD) using Magnetic Resonance Imaging (MRI).
Patients and Methods: We prospectively studied 30 pancreatic insufficient CF patients, mean age 25.9 (17–42 years). Diagnosis of CF was established by clinical symptoms, sweat tests and presence of known CF mutations. 14 patients had CFRD and 16 patients were non diabetic according to World Health Organization criteria. None of the non diabetics had impaired oral glucose tolerance tests and CFRD patients were receiving insulin replacement therapy at the time of the study. MRI scans were acquired using T1 weighted fat suppression sequences and assessed by an examiner (MC) unaware of patients' diabetic status.
Results: The 2 groups were matched for age and gender. There were no differences in body mass index or exocrine pancreatic function as assessed by the requirement of oral lipase. CFRD patients had lower FEV1 and FVC, but this did not attain statistical significance. The pancreas was visible by MRI in only 4 of 14 (28.6%) patients with CFRD and in 5 of 16 (31.3%) patients without CFRD. In a total of 21 of 30 (70%) patients with and without CFRD the pancreas was not detected by MRI.
Conclusions: This is the first study to show that the pancreas is not measurable as an entity in the majority of CF patients irrespective of their diabetic status. The fact that the pancreas was not visible by MRI even in patients without clinical or biochemical evidence of diabetes raises the question as to why not all patients with non visible pancreas are diabetics.
Supported by: The Charitable Trustees, United Bristol NHS Trust, UK.

330 Acid steatocrit is not helpful in cystic fibrosis patients with mild or no steatorrhea

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The applicability of acid steatocrit (AS) in the assessment of fecal fat losses has been assessed with contradictory results. In the present study, we have aimed to determine the role of AS in in CF patients with mild (<10g/d) or no steatorrhea.
Material & methods: The study comprised 45 CF patients aged 7 to 18 years. In all subjects fecal fat concentration (FFC) and excretion (FFE) in day stool collection was assessed as well as AS values in a single stool sample were determined.
Results: 130 triple results (FFC, FFE, AS) were available for the analysis. The range of values (median; mean±SEM) of FFE, FFC and AS were as follow: 0.8–9.9 g/day (4.2%; 4.9±0.2), 0.7–30.6 g/g (4.0; 6.2±0.6) and 4.0–35.0% (10.0; 13.1±0.7). AS correlated both with FFE ($r < 0.203$, $p < 0.021$) and FFC values ($r < 0.224$, $p < 0.01$). The correlations, although statistically significant, were very weak. The specificity and sensitivity of AS in the determination of abnormal FFC were low: 59.2% and 54.0% for the cut-off level of 10% and 31.5% and 84.2% for the cut-off level of 20%. Similarly, low PPV and NPV values were observed (47.8% and 65.1% for a lower cut-off level and 58.6% and 63.4% for a higher cut-off level).
Conclusions: Acid steatocrit does not reflect fecal fat excretion in cystic fibrosis patients with mild or no steatorrhea. Moreover, its applicability in the assessment of fecal fat concentration in this subgroup of patients has no practical value.
Supported by: Grant from The Poznan University of Medical Sciences.

331 Adiponectin may reflect the degree of the inflammatory, fibrosing process in the cystic fibrosis liver

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Steatosis and an inflammatory, reactive-fibrosing process leads to focal and multilobular biliary cirrhosis in cystic fibrosis (CF). Serum adiponectin (AD) is elevated in CF patients.
Aim: To investigate any correlation between the degree of liver involvement and serum AD.
Methods: Hepatic ultrasound was performed in 21 CF patients and 10 controls. Liver involvement was demonstrated in 12 CF patients, who underwent percutaneous liver biopsy with the biopsy needle directed to the site of the lesion with CT scan. The grading scale of CF-related liver histology was: 1 = normal, 2 = steatosis, 3 = steatosis + inflammatory reaction, 4 = inflammatory reaction + fibrosis (focal biliary cirrhosis) and 5 = multilobular biliary cirrhosis. Five groups were studied: Group A = Controls (10, mean age 13.50 yrs); Group B = CF patients having no liver disease (9, mean age 20 years); Group C = 4 CF patients – grade 2 or 3; Group D = 4 CF patients – grade 4; and Group E = 4 CF patients – grade 5. The mean age of CF-LD patients was 14.50 yrs.
Results: AD was significantly different ($p = 0.005$) between CF patients having liver involvement (mean AD 10.59 μg/ml) and those having normal liver (mean AD 7.36 μg/ml) as well as controls (mean AD 5.61 μg/ml). Furthermore, significant correlation was demonstrated (one way ANOVA, $p = 0.000$) between the grading of liver involvement and AD (control = 5.61, grade 1 = 7.36, grade 2+3 = 7.41, grade 4 = 8.38, grade 5 = 15.49 (μg/ml).
Conclusion: Adiponectin measurement may be a screening test for the assessment of the degree of the inflammatory, reactive fibrosing process in the liver of CF patients.